PUBLICATIONS BY ISPGHAN MEMBERS IN PUBMED INDEXED JOURNALS

(1st January, 2022 – 15th March, 2022)
Updated as on 15th March, 2022

JANUARY 2022

[1.] Jagadisan B, Dhawan A. Emergencies in Paediatric Hepatology. J Hepatol. 2022 Jan 3:S0168-8278(21)02304-7. doi: 10.1016/j.jhep.2021.12.027. Epub ahead of print. PMID: 34990749.

In this review article the authors have chosen to address 5 emergencies in pediatric hepatology: infants with liver disease; acute liver failure; management of bleeding varices; liver-based metabolic disorders; and liver tumours and trauma. A wide spectrum of conditions that cause liver disease in infants may present as conjugated jaundice, which could be the only symptom of time sensitive disorders wherein algorithmic multistage testing is required for accurate and early diagnosis, while vitamin K, specific milk formulae and disease-specific medications are essential to avoid mortality and long-term morbidity. Management of paediatric acute liver failure requires co-ordination with a liver transplant centre, safe transport and detailed age-specific aetiological work-up – clinical stabilisation with appropriate supportive care is central to survival if transplantation is indicated. Gastrointestinal bleeding may present as the initial manifestation or during follow-up in patients with portal vein thrombosis or chronic liver disease and can be managed by pharmacotherapy with octreotide, or endotherapy (variceal ligation/sclerotherapy/glue injection) or radiological interventions [transjugular intrahepatic portosystemic shunt (TIPS)]. Liver-based inborn errors of metabolism may present as encephalopathy that needs to be recognised and treated early to avoid further neurological sequelae and death. Liver tumours and liver trauma are both rare occurrences in children and are best managed by a multidisciplinary team in a specialist centre.

[2.] Mehta SJ, Malhotra S, Panwar A, Sibal A. Complicated Pylephlebitis Secondary to Perforated Appendicitis in an Adolescent. J Indian Assoc Pediatr Surg. 2022 Jan-Feb; 27(1):115–117.

In this case report authors have presented a case of pylephlebitis secondary to perforated appendicitis with incomplete resolution of portal vein thrombosis in a 12 year old boy who presented with high grade fever with chills and abdominal pain for 10 days. He underwent laparotomy and appendectomy followed by prolonged anticoagulation. Authors have shown that septic pylephlebitis in a setting of acute appendicitis requires a high index of suspicion and careful evaluation and follow-up is essential in children with added complications.

[3.] Wadhwa A, Kesavelu D, Kumar K, Chatterjee P, Jog P, Gopalan S, Paul R, Veligandla KC, Mehta S, Mane A, Pandit S, Rathod R, Jayan S, Mitra M. Role of Lactobacillus reuteri DSM 17938 on Crying Time Reduction in Infantile Colic and Its Impact on Maternal Depression: A Real-Life Clinic-Based Study. Clin Pract. 2022 Jan 7; 12(1):37–45.

In this study authors have evaluated the role of L. reuteri DSM 17938 in infantile colic in India. They have observed that L. reuteri DSM 17938 supplementation in infantile colic subjects resulted in a significant reduction of crying time and unexplained fussiness. A significant number of subjects reported 50% reduction in crying time throughout the study duration. There was also a significant reduction in maternal depression scores, from baseline to study end.

[4.] Saini K, Bolia R, Bhat NK. Incidence, Predictors and Outcome of Sepsis-Associated Liver Injury in Children: A Prospective Observational Study. Eur J Pediatr. 2022 Jan 12:1–9. doi: 10.1007/s00431-022-04374-2. Epub ahead of print. PMID: 35020050.

In this prospective observational cohort study authors have enrolled 127 children < 19 years with diagnosis of sepsis over a period of 12 months. Clinical and biochemical parameters of children with sepsis who developed sepsis-associated liver injury [SALI] were compared with those without SALI to determine the risk factors of SALI and its impact on in-hospital mortality. The SALI pattern was cholestatic in 18 (40%), hepatocellular in 17 (37.7%) and hypoxic hepatitis in 10 (22.3%). They have concluded that SALI develops in 45 (31.3%) with sepsis. A higher pSOFA score [>9.5] is associated with SALI. Children who develop SALI have a twofold higher risk of mortality than those without SALI.

[5.] Prasad D, Sen Sarma M, Yachha SK, Prasad R, Srivastava A, Poddar U, Kumar A. Can we Predict Early Renal Impairment in Pediatric Cirrhosis? Indian J Gastroenterol. 2022 Jan 24. doi: 10.1007/s12664-021-01190-8. Epub ahead of print. PMID: 35067841.

In this study authors have aimed to detect early renal dysfunction (RD) in cirrhotic children by renal resistive index (RI) and plasma aldosterone (PA) and evaluated the effects of large-volume paracentesis (LVP) and albumin infusion on the same. 99 non-azotemic cirrhotic children with tense ascites (undergoing LVP with albumin infusion) were prospectively enrolled. Blood biochemistry and doppler ultrasonography for RI and PA were measured at regular intervals. RI >0.7 was considered as RD. Outcomes were noted at D90 and 1 year. Chronic liver disease children without ascites were included as controls. Baseline RD was observed in 32% and was significantly higher in tense ascites compared to controls. Tense ascites with RD at admission had higher chances of acute kidney injury (AKI), ascites recurrence, hospital readmission, and mortality compared to patients without RD by D90. Significant reduction in RI was noted at 48 h, D7, D30, and

D90 compared to baseline after LVP with albumin. Pediatric End-stage Liver Disease (PELD) score and PA had a strong positive correlation with baseline RI. Using multivariate analysis, PELD score and PA were predictors of AKI and mortality. Authors have concluded that abnormal baseline RI can be used as an early predictor of RD and predict long-term renal outcomes in pediatric cirrhosis. Baseline RI correlated well with the severity of liver disease and PA. Paracentesis and albumin infusion effectively reduced RI.

[6.] Alam S, Lal BB. Recent Updates on Progressive Familial Intrahepatic Cholestasis Types 1, 2 and 3: Outcome and Therapeutic Strategies. World J Hepatol. 2022 Jan 27; 14(1):98–118.

In this review article authors have described the recent advances in understanding the clinical course of progressive familial intrahepatic cholestasis types 1, 2 and 3 and the role of emerging therapies. They have highlighted that genotype correlates well with phenotype in PFIC2 but not in PFIC1. The expanded role of the heterozygous transporter defects presenting late needs to be suspected and identified even in adulthood. Medical therapy and surgical biliary diversion (SBD) form the cornerstone of the management of pruritus. Liver transplantation in these children is associated with unique issues like a high rate of intractable diarrhoea, growth failure, steatohepatitis and graft failure in PFIC1 and antibody-mediated BSEP deficiency in PFIC2. There is a promising role of apical sodium-dependent bile acid transporter (ASBT) inhibitors in the management of cholestasis.

[7.] Ravindranath A, Sarma MS. Mitochondrial Hepatopathy: Anticipated Difficulties in Management of Fatty Acid Oxidation Defects and Urea Cycle Defects. World J Hepatol. 2022 Jan 27; 14(1):180–194.

In this minireview authors have elaborated on the dilemmas in the diagnosis and management of fatty acid oxidation defect (FAOD) and urea cycle defects (UCD). The cornerstone in the management of FAOD is avoiding hypoglycaemia. They have discussed the role of carnitine supplementation, dietary interventions, newer therapies like triheptanoin, long-term treatment and approach to positive newborn screening. In UCD the general goal is to avoid excessive protein intake and indigenous protein breakdown. They have highlighted that striking the right balance between avoiding hyperammonemia and preventing deficiencies of essential nutrients is a formidable task. Practical issues during the acute presentation including differential diagnosis of hyperammonemia, dietary dilemmas, the role of liver transplantation, management of the asymptomatic individual and monitoring have been elaborately discussed.

FEBRUARY 2022

[8.] Karri PS, Jagadisan B, Lakshminarayanan S, Plakkal N. Biliary Atresia Screening in India Strategies and Challenges in Implementation. Indian J Pediatr. 2022 Feb; 89(2):133–140.

In this clinical trial authors have aimed to assess the utilization of stool color card (SCC) for biliary atresia (BA) screening implemented in southern India and the effect on SCC utilization of face-to-face verbal education versus video-based content delivery. The study included newborns with postnatal age of less than 2 wk at discharge. Mothers were administered SCCs and provided standardized verbal or video health education based on the time-period of enrollment. Home based monitoring of stool color and return of SCC on postnatal day 21 was advised. Telephone surveys were conducted to identify SCC use among families that did not return the SCC by post. Of the 2254 newborns enrolled, 1130 were in the verbal-counseling group and 1124 in the video-counseling group. No newborns with pale stools and biliary atresia were identified. SCC return rates were 3.8% and 2.8%. Comparing the verbal and video-counseling groups, there were no differences in the conservative (81.8% vs. 81.5%) and optimistic estimates (97.1% vs. 97.3%) of SCC utilization rates. Mothers with better educational status had higher optimistic estimates of SCC utilization. Authors have concluded that good utilization rates of SCC with video content delivery of health information thereby demonstrating a scalable model for BA screening in the southern Indian population.

[9.] Menon J, Shanmugam N, Vij M, Rammohan A, Rela M. Multidisciplinary Management of Alagille Syndrome. J Multidiscip Healthc. 2022 Feb 23; 15:353–364. doi: 10.2147/JMDH.S295441. PMID: 35237041.

In this review article authors have stressed on the need for multidisciplinary management of Alagille syndrome (ALGS) given the systemic manifestations of ALGS are protean and a wider spectrum is being increasingly elucidated. ALGS is usually suspected in infants presenting with high gamma glutamyl transpeptidase cholestasis and/or congenital heart disease. In children it may present with decompensated cirrhosis, intellectual disability or short stature, and in adults vascular events like stroke or ruptured berry aneurysm are more commonly noted. Liver transplantation (LT) is indicated in children with cholestasis progressing to cirrhosis with decompensation, intractable pruritus, recurrent fractures, hepatocellular carcinoma and disfiguring xanthomas. Due to an increased risk of renal impairment noted in ALGS, these patients would require optimized renal sparing immunosuppression in the post-transplant period. Cardiovascular involvement is an important cause of morbidity and mortality not only in children but also in the adult patients suffering from ALGS and needs to be recognized and treated as early as possible.

MARCH 2022

[10.] Ganesh R, Sathiyasekeran M, Srinivas S, Narayanan RK. Clinical Spectrum of Monogenic Infantile-Onset Inflammatory Bowel Disease. Indian J Pediatr. 2022 Mar 5. doi: 10.1007/s12098-022-04103-5. Epub ahead of print. PMID: 35246832.

In this clinical brief the authors have collated the clinic-laboratory profile, molecular genetic testing and treatment details of 8 children diagnosed with monogenic infantile-onset IBD. The median age at onset of symptoms was 3 mo. Sibling death and consanguinity were noted in 50% each respectively. Diarrhea was the presentation in 100% and hematochezia in 62%. Colonic ulcers on colonosopy was seen in 7 infants. The common mutation identified was IL-10R gene in 3 (42%) and LRBA gene mutation in 2 (25%). HSCT was done in 4 children and the rest were managed conservatively. Although there was no mortality in this series, two children (25%) were lost for follow-up. This preliminary study highlights a spectrum of monogenic variants seen in IOIBD and emphasizes the importance of early diagnosis for definitive therapy.

MISSED INADVERTENTLY IN PREVIOUS ISSUES 2021

[1.] Krishnan, Mouleeswaran, Gopal et al. First Report of a Paediatric Collision Tumour in the Liver Recognised After Liver Transplantation: Blissful Ignorance Has Benefits!, Journal of Clinical and Experimental Hepatology, https://doi.org/10.1016/j.jceh.2021.05.006. Epub ahead of print.

Simultaneous development of two discrete liver tumours of distinct histologies (collision tumour) has been occasionally reported in adults but never in children. The authors have presented the first reported case of hepatic collision tumours (hepatocellular carcinoma and cholangiocarcinoma) in the explant liver of a 12 year old child who underwent living donor liver transplantation for end-stage liver disease and severe hepatopulmonary syndrome

[2.] Hosaagrahara Ramakrishna et al., When Push Comes to Shove! Emergency ABO-Incompatible Pediatric Living Donor Liver Transplant for Acute Wilson's Disease, Journal of Clinical and Experimental Hepatology, https://doi.org/10.1016/j.jceh.2021.08.008. Epub ahead of print.

In this case report authors have reported on 8 year old girl with acute Wilson's disease with rapidly deteriorating liver function who underwent a successful ABO-incompatible living donor liver transplant (ABOi-LDLT) using a rapid desensitization protocol which included a single dose of Rituximab along with mycophenolate mofetil (MMF) and standard plasma exchange.

[3.] Krishnan SK, Varghese J, Patcha R, Hosaagrahara Ramakrishna S, Reddy MS. Delayed Expulsion of biliary cast after hepaticojejunostomy for post-liver transplantation biliary stricture-better cut than canulate? Pediatr Transplant. 2021 Dec; 25(8):e14115. doi: 10.1111/petr.14115. Epub 2021 Aug 19. PMID: 34409710.

In this case report authors have described performing Roux en Y hepaticojejunostomy to treat an anastomotic biliary stricture with biliary cast formation which developed in a 1 year old child who underwent living donor liver transplantation (LDLT) for acute liver failure. Two months after the surgery, the child passed a large biliary cast in the stools. This reiterates the advantage of wide biliary drainage provided through surgical therapy even though endoscopic therapy is usually the first choice of treatment.

[4.] Pratim Nayak, S., Hosaagrahara Ramakrishna, S., Jyothinagaram Sivaprakas, B. et al. Mauriac syndrome: a rare cause of massive hepatomegaly. Int J Diabetes Dev Ctries 41, 697–699 (2021). https://doi.org/10.1007/s13410-021-00939-3.

Authors have reported a 10-year-old boy with poorly controlled type I diabetes mellitus (on pre-mix insulin), hepatomegaly, and extremely high levels of liver transaminases. After exclusion of other causes of hepatitis, his blood sugars were controlled using basal bolus insulin regimen and ensuring good compliance to treatment. In an about 8 weeks, his transaminases normalized and hepatomegaly regressed. This case report illustrates Mauriac syndrome or glycogen hepatopathy (GH) which is an uncommon complication of poorly controlled type 1 diabetes mellitus commonly occurring in adolescents.

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CORRIGENDUM

This is the correct citation of the first article published under September 2021 in the last issue

Poddar U, Benjamin M, Aggarwal R, Sarangi AN, Mathias A, Yachha SK, Srivastava A. Hepatitis B Transmission: Is Vertical Transmission the Major Route in Intermediate Endemic Areas? A Proof-of-Concept Study Based on Mother-Child Genotypes. Am J Trop Med Hyg. 2021 Sep 27; 105(6):1569-1574.

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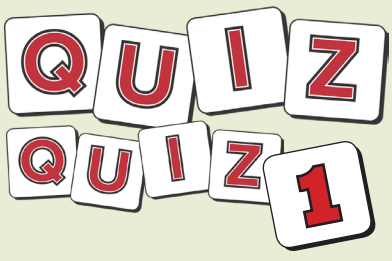
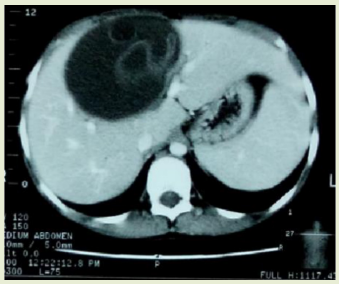
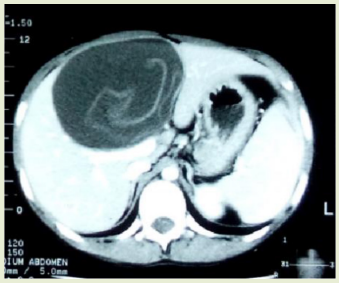


A 14 year old girl presented with progressively enlarging lump and pain in right hypochondrium with intermittent fever for last 3 years. Complete blood picture and liver function tests were largely unremarkable. A contrast enhanced computerised tomography was performed.

What is the diagnosis and possible modalities of management (in brief)?





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